CHES STUDIES

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1988-1990 NORTH CAROLINA BIRTH DEFECTS REGISTRY REPORT

by

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ABSTRACT

This report presents data from the 1988-90 North Carolina Birth Defects Registry. Data tables within this report are updated to include the 1989 and 1990 Registry data, and in some cases overlap the 1988 data tabulated in a previous CHES study on birth defects[1]. Descriptive statistics displayed in the tables give the reader perspectives on the prevalence of birth defects in the state. Other sections of this report are written with the intention of orienting readers to the use and interpretation of data on birth malformations. The report also discusses potential problems associated with analyzing rates based on small numbers, and the effects of variations in data collection methods on geographic and temporal patterns of congenital malformations in the state.

Because one of the Healthy Carolinians objectives is to reduce the state's infant death rate to seven deaths per 1,000 live births by the Year 2000[2], the study of birth defects as they relate to infant deaths has gained importance. From 1988 through 1990 birth defects were the leading cause of infant mortality in North Carolina. In 1990, 19.3 percent of the infant deaths were due to congenital anomalies.

DATA SOURCES FOR CONGENITAL ANOMALIES

The North Carolina Birth Defects Registry uses a multi-source case ascertainment system to identify infants with birth defects. The data systems currently used in the registry are birth and infant death certificates, hospital discharge records, Medicaid claims, Children's Special Health Services (CSHS) records and neonatal intensive care unit (NICU) reports.

Table 1 shows, for each data source, the number and percentage of all live births with reported congenital malformations. The increase in the number of Medicaid infants with birth defects probably reflects the increase in the percentage of all deliveries paid for by Medicaid during this period, which rose from 23.5 percent in 1988 to 35.6 percent in 1990.[3] The increase in 1989 in the number of hospital discharge records indicating congenital anomalies may be due, in part, to more complete discharge data provided by the Medical Database Commission.

As more data sources are linked together, an increasing number of records are composed of information from more than one data set. With the addition of the NICU discharge and statewide hospital discharge data sources, records with multiple sources of data increased from 25.2 percent in 1988 to 36.8 percent in 1990 (Table 2). The percentage of cases uniquely ascertained from the birth certificate, infant death certificate, and Medicaid claims has declined since 1988. Since both the Medicaid and hospital discharge files are billing records, most of the Medicaid claims are duplicated in the hospital discharge file beginning in 1989. The total number of records with reported congenital anomalies in 1990 was 6,465, an 11 percent increase over 1989.

Table 1

Numbers and Percentages of All Congenital Anomalies (ICD-9 740-759) Reported by Source of Data North Carolina, 1988-90

	Number of Records with Reported Congenital Anomalies			Percent of All Births*		
Source of Data	1988	1989	1990	1988	1989	1990
Birth Certificates	1,043	1,001	1,113	1.1	1.0	1.0
Medicaid Claims	1,189	1,465	1,995	1.2	1.4	1.9
Infant Deaths	256	232	247	0.3	0.2	0.2
Hospital Discharge	2,488	4,092	4,745	2.6	4.0	4.5
CSHS	740	994	906	0.8	1.0	0.9
NICU	N/A	504	503	N/A	0.5	0.5

*Total number of resident live births:
1988 = 97,560
1989 = 102,091
1990 = 104,439

Table 2

Numbers and Percentages of All Congenital Anomalies (ICD9 740-759) Reported by Unique and Multiple Data Sources North Carolina, 1988-90

Source of Data	Number of Records			Percent of all Records with Congenital Anomalies		
	1988	1989	1990	1988	1989	1990
Sole Source						
Birth Certificates	517	341	441	11.7	5.9	6.8
Medicaid Claims	490	113	106	11.1	1.9	1.6
Infant Deaths	92	78	90	2.1	1.3	1.4
Hospital Discharge	1,530	2,267	2,418	34.6	39.0	37.4
CSHS	675	900	812	15.3	15.5	12.6
NICU	N/A	229	221	N/A	3.9	3.4
Multiple Sources	1,116	1,887	2,377	25.2	32.5	36.8
Total	4,420	5,815	6,465	100	100	100

DISTRIBUTION OF ANOMALIES BY ORGAN SYSTEM AND CASE CHARACTERISTICS

In **Table 3**, rates for selected birth defects and birth conditions are listed for 1988, 1989 and 1990. The rates represent those infants with Registry records that have one or more of the (listed) diagnoses. Children with two or more heart malformations, for instance, would be included only once in the heart malformations total rate. The same applies to categories such as "other musculoskeletal/integumental anomalies". At the bottom of **Table 3**, the rates for all infants having birth defects in 1988, 1989, and 1990 are shown.

Rates for some birth defects, such as anencephalus, anophthalmos/microphthalmos, and coagulation defects have decreased over the period 1988-90. However, the small number of cases precludes meaningful interpretation of these apparent "trends". Rates for other malformations, such as hydrocephalus, tetralogy of Fallot, omphalocele/gastroschisis, Down syndrome, and congenital syphilis, have increased over the period. The increases in the rates of some congenital malformations from 1988 to 1989 may be due, in part, to improved ascertainment of these conditions through the NICU and statewide hospital discharge data. Changes from 1989-90 were probably not due to changes in data sources, but may still reflect differences in case ascertainment within a particular data source, or they may be attributed to random fluctuations.

Table 3

Rates* for Selected Birth Defects
1988-90 North Carolina Birth Defects Registry

	RATE			
	1988	1989	1990	1988-90
CENTRAL NERVOUS SYSTEM				
Anencephalus (740.0)	1.6	0.9	1.0	1.2
Spina Bifida (741.0, 741.9)	4.6	3.5	5.2	4.4
Encephalocele (742.0)	1.2	1.2	0.9	1.1
Hydrocephalus (742.3)	5.7	7.1	8.1	7.0
Microcephalus (742.1)	2.6	2.9	2.8	2.8
Other Central Nervous System Anomalies (742.2, 742.4-.9)	3.4	4.0	4.0	3.8
EYE				
Anophthalmos/Microphthalmos (743.0, 743.1)	0.8	0.6	0.3	0.6
Congenital Cataract (743.3)	0.4	1.1	1.1	0.9
CARDIOVASCULAR				
Heart Malformations (745.0-746.9)	41.3	51.4	51.2	48.1
Common Truncus (745.0)	0.0	0.2	0.8	0.3
Transposition of Great Arteries (745.1)	1.2	2.1	2.1	1.8
Tetralogy of Fallot (745.2)	0.7	2.0	1.8	1.5
Ventricular Septal Defect (745.3,.4,.7)	9.5	15.7	14.5	13.3
Atrial Septal Defect (745.5)	2.5	5.6	3.1	3.7
Endocardial Cushion Defect (745.6)	0.7	0.9	1.8	1.2
Anomalies of Pulmonary Valve (746.0)	1.6	7.1	5.7	4.9
Tricuspid Valve Atresia and Stenosis (746.1)	0.1	0.5	0.3	0.3
Aortic Valve Stenosis and Atresia (746.3,.4)	0.5	1.3	0.6	0.8
Hypoplastic Left Heart Syndrome (746.7)	2.3	2.4	2.1	2.2
Other Circulatory/Respiratory Anomalies (747.0-748.9)	39.8	61.0	58.2	53.2
Patent Ductus Arteriosus (747.0)	19.8	32.3	30.4	27.7
Coarctation of Aorta (747.1)	0.3	2.1	1.9	1.4
Pulmonary Artery Anomaly (747.3)	1.2	2.3	2.0	1.8
Agenesis of Lung (748.5)	6.0	7.1	8.0	7.0
OROFACIAL				
Cleft Palate without Cleft Lip (749.0)	3.6	3.2	3.9	3.6
Cleft Lip with and without Cleft Palate (749.1,.2)	4.3	9.3	5.8	6.5
GASTROINTESTINAL				
Rectal Atresia/Stenosis (751.2)	3.2	2.9	3.5	3.2
Tracheo-esophageal Fistula/Esophageal Atresia (750.3)	1.6	2.9	3.0	2.5
Other Gastrointestinal Anomalies (750.0-.2, 750.4-.9, 751.0,.1, 751.3-.9)	9.3	15.0	16.3	13.6

Table 3 (continued)

Rates* for Selected Birth Defects
1988-90 North Carolina Birth Defects Registry

	RATE			
	1988	1989	1990	1988-90
GENITOURINARY				
Malformed Genitalia (752.0-752.9)	57.6	66.8	69.3	64.9
Renal Agenesis (753.0)	2.4	2.6	1.9	2.3
Other Urogenital Anomalies (753.1-.9)	8.1	10.8	13.8	11.0
MUSCULOSKELETAL				
Polydactyly/Syndactyly (755.0,.1)	38.2	46.3	47.3	44.1
Clubfoot (754.5-.7)	44.5	55.6	50.2	50.2
Omphalocele/Gastroschisis (756.7)	5.8	7.5	12.3	8.6
Diaphragmatic Hernia (756.6)	4.2	3.5	4.7	4.1
Other Musculoskeletal/Integumental Anomalies (754.0-.4, 754.8, 755.2-.9, 756.0-.5, 756.8-.9, 757.0-.9)	179.8	231.5	275.6	230.0
CHROMOSOMAL				
Down Syndrome (758.0)	6.7	8.2	9.5	8.2
Other Chromosomal Anomalies (758.1-.9)	4.5	5.4	5.4	5.1
Trisomy 13 (758.1)	0.8	1.6	1.0	1.1
Trisomy 18 (758.2)	1.6	1.1	1.7	1.5
INFECTIONS				
Congenital Syphilis (090.0-090.9)	1.1	3.0	4.8	3.0
Gonococcal Infections (098.0-098.8)	0.3	0.2	0.1	0.2
METABOLIC DISORDERS				
Cystic Fibrosis (277.0)	0.4	0.9	0.5	0.6
Phenylketonuria (270.1)	0.5	0.0	0.0	0.2
OTHER CENTRAL NERVOUS SYSTEM DISORDERS				
Infantile Cerebral Palsy (343.0-343.9)	4.0	5.6	5.1	4.9
Epilepsy (345.0-345.9)	1.1	1.8	1.5	1.5
OTHER CONDITIONS				
Congenital Hypothyroidism (243)	0.5	0.7	0.5	0.6
Coagulation Defects (286.0-286.9)	1.4	0.8	0.9	1.0
Fetal Alcohol Syndrome (760.71)	4.8	5.7	5.9	5.5
Hereditary Hemolytic Anemias (282.0-282.9)	2.3	2.1	1.9	2.1
Total Number of Live Births	97,560	102,091	104,439	304,090
Overall rate of birth defect occurrence	540.4	701.0	776.3	675.4

* per 10,000 live births

Table 4 shows the percentages of infants with selected maternal and perinatal characteristics who were born with congenital anomalies (ICD-9 740-759). Of all infants whose mothers were less than 18 years of age in 1990, 8.4 percent were born with major and/or minor congenital anomalies. A higher percentage of male infants than females were born with birth defects for all three years. The frequency of reported congenital malformations among nonwhites was twice that of whites throughout the period. Most, but not all, of the difference in the prevalence of congenital malformations between whites and nonwhites is due to a disproportionate number of minor malformations being reported among blacks.

Table 4

Percentage of Births with One or More Congenital Anomalies* by Selected Maternal and Perinatal Characteristics North Carolina, 1988-90

Maternal and Perinatal Characteristics		Percentage of Births With a Congenital Anomaly		
		1988	1989	1990
Age of Mother	Less than 18 years	6.8	7.2	8.4
	18-34 years	4.4	5.6	6.0
	35 years and over	4.3	5.6	6.3
Gender of Child	Male	5.0	6.2	6.7
	Female	4.1	5.2	5.6
Race	White	3.3	4.3	4.5
	Nonwhite	7.1	8.7	9.8
Education of Mother	Less than 9 years	6.0	7.0	8.1
	9-11 years	6.1	6.9	7.7
	12 years	4.6	6.0	6.3
	More than 12 years	3.4	4.5	5.1
Birthweight	Less than 1,500 grams	13.0	18.4	21.1
	1,500-2,499 grams	7.7	10.3	10.8
	2,500 grams and over	4.2	5.1	5.6
Infant Death	Yes	27.0	29.2	29.0
	No	4.3	5.4	5.9
Medical Risk Factors	Yes	5.6	7.0	7.7
	No	4.2	5.3	5.7
Smoked During Pregnancy	Yes	4.8	6.0	6.5
	No	4.6	5.6	6.1
C-Section	Yes	5.4	6.7	7.4
	No	4.3	5.4	5.8

* Includes both major and minor malformations

Very low birthweight (less than 1,500 grams) and intermediate low birthweight (1,500-2,499 grams) babies were born with congenital anomalies more often than their counterparts weighing 2,500 grams or more. In 1990, 21.1 percent of the very low birthweight infants were born with a congenital abnormality. In comparison, only 5.6 percent of all babies weighing over 2,500 grams were reported to have congenital anomalies in 1990. The increased likelihood of patent ductus arteriosus and lung hypoplasia associated with preterm infants probably accounts for much of the difference between very low birthweight and normal birthweight infants.

The number and percentage of infants in the state who died within their first year of life, and who had been diagnosed with a congenital anomaly, increased from 1988 to 1989. In 1988 and 1990 respectively, 27 percent and 29 percent of all infants who died within the first year of life had been diagnosed with one or more congenital anomalies. In about one-third of these cases, however, birth defects were not reported to be the underlying cause of death. The percentage of infants who lived through their first year of life and had at least one congenital anomaly increased from 4.3 in 1988 to 5.9 in 1990. Again, part of this increase is due to better ascertainment of congenital anomalies beginning in 1989.

CAUTIONS IN DATA INTERPRETATION

The Registry data prior to 1989 have some limitations. One limitation is the incomplete hospital discharge data in the 1984-88 Registry files. Because of the "pockets" of missing hospital discharge data in certain parts of the state, the Registry data prior to 1989 may underestimate the occurrence of certain types of congenital malformations in particular regions or counties of North Carolina. Since the newborn hospital discharge data is a major contributor of infant birth defect data, it has been useful to determine where the "pockets" of missing data are located. Figure 1 shows the counties that had complete birth defect reporting, partial reporting, and no reporting in 1988.

A second limitation is the under-reporting or incompleteness of birth defects data. For example, the birth certificate and newborn hospital discharge databases record birth defect diagnoses made during the newborn period only, and are poor sources of data for conditions that are generally not diagnosed until after the infant is discharged from the hospital of birth.

Thirdly, the hospital discharge data are primarily collected for insurance reimbursement and other purposes, and are not coded in a format that is most desirable for etiologic studies. Hospital discharge summaries record up to five diagnoses, so if an infant has more than five birth defects and/or other health conditions, some diagnoses are lost when the original hospital record data are transferred into the hospital discharge database format. Despite these data limitations, ascertainment of many of the more severe malformations, such as spina bifida, renal agenesis, and hypoplastic left heart syndrome, appears to be relatively good. The rates for these defects are comparable to those reported by the National Birth Defects Monitoring Program.

When studying a public health problem, such as the occurrence of birth defects, counts and rates are used as tools for evaluating the extent of the problem. An incidence rate measures the occurrence of new cases of disease in a community, and is useful in helping to determine the need for initiation of preventive measures. "The term rate, although there are some exceptions, is usually reserved to refer to those calculations that imply the probability of the occurrence of some event".[4] Any rate with a small number of events in the numerator will be unstable, with possibly large random fluctuations from year to year that do not comprise a significant trend. It has been shown that events of a rare nature follow a Poisson probability distribution. A useful rule of thumb is that any rate based on fewer than 20

events in the numerator may have a 95 percent confidence interval that is wider than the rate itself.[5] Caution should be used when interpreting those rates in **Table 3** that are less than 2.0, as these rates generally reflect fewer than 20 observed cases.

Since the North Carolina Birth Defects Registry uses a passive surveillance system with secondary data sources, the state data will tend to underestimate the actual occurrence of birth defects. Unlike cancer, birth defect reporting from hospitals or physicians in North Carolina is not mandatory by law.

CONCLUSIONS

The primary goal of the North Carolina Birth Defects Registry is to collect and analyze data on infants with birth defects. The Registry's purpose is to provide health care providers and policymakers with information necessary to plan, develop, and implement strategies for the treatment and prevention of serious congenital malformations. The State Center for Health and Environmental Statistics is currently evaluating the utility of the Registry for better serving the needs of the public and the medical community. We welcome your comments or suggestions.

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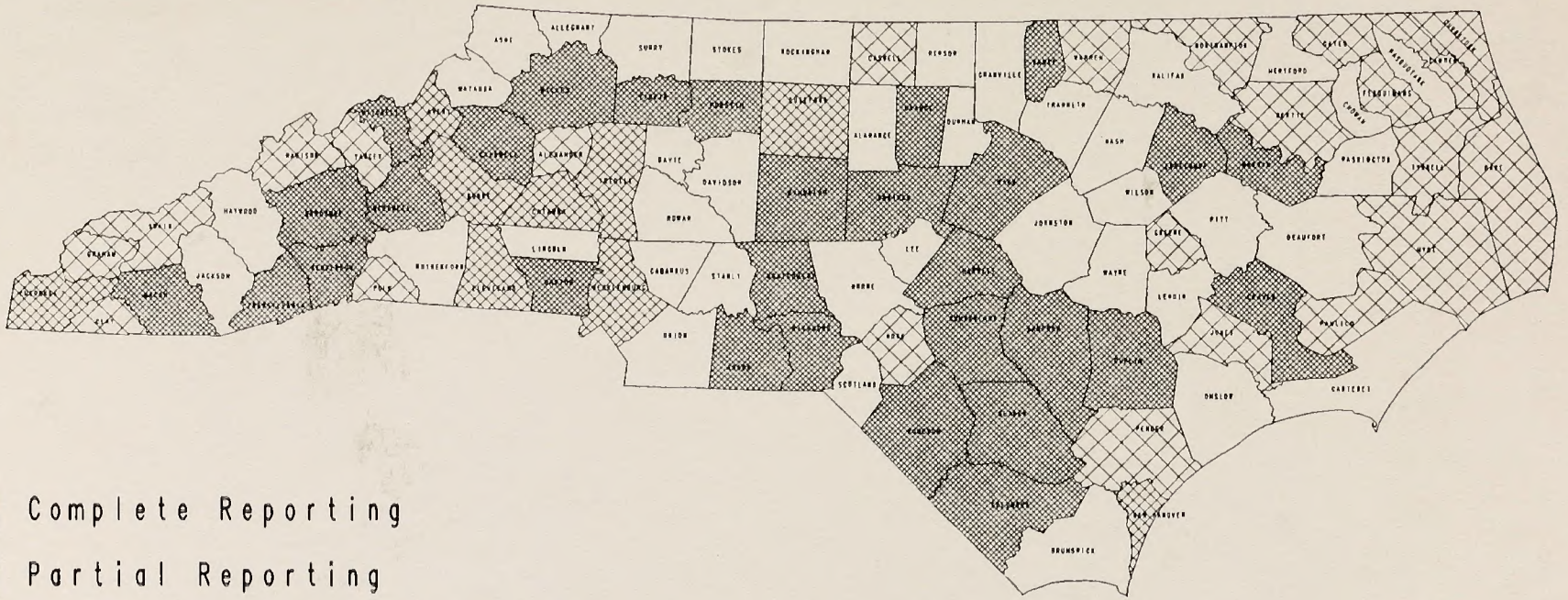
Figure 1

1988 COUNTY NEWBORN HOSPITAL DISCHARGE REPORTING STATUS FOR INFANTS HAVING BIRTH DEFECTS



- Complete Reporting
- Partial Reporting
- No Birthing Hospitals
- No Reporting

Source: North Carolina Birth Defects Registry

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